

Supplementary Table S6.

Data extracted for the “Prognostic Outcome” domain and its variables, derived from 36 papers.

Prognostic outcome	N° papers	references	N° patients	references
MYOCARDITIS	1	(1)	2 / 46 (4 %)	from 1 paper (1)
HEART FAILURE	23	(2-24)	138 / 1814 (8 %)	from 15 papers (2, 4, 5, 10-12, 14, 15, 17-22, 24)
ACUTE CORONARY SYNDROME	7	(2, 4, 5, 20, 25-27)	29 / 687 (4 %)	from 7 papers (2-5, 20, 25, 27)
SIGNIFICANT ARRHYTHMIAS	18	(2-5, 9, 10, 12, 17, 18, 20, 24-26, 28-32)	197 / 3106 (6 %)	from 16 papers (2-5, 9, 10, 12, 17, 18, 20, 24, 25, 28, 29, 31, 32)
SUDDEN DEATH	11	(4, 8, 10, 19, 26, 27, 31, 33-36)	119 / 1063 (11 %)	from 10 papers (4, 8, 10, 19, 26, 27, 31, 33, 35, 36)

REFERENCES

1. Mavrogeni SI, Bratis K, Karabela G, Spiliotis G, Wijk K, Hautemann D, et al. Cardiovascular Magnetic Resonance Imaging clarifies cardiac pathophysiology in early, asymptomatic diffuse systemic sclerosis. *Inflamm Allergy Drug Targets.* 2015;14(1):29-36.
2. Tyndall AJ, Bannert B, Vonk M, Airo P, Cozzi F, Carreira PE, et al. Causes and risk factors for death in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Ann Rheum Dis.* 2010;69(10):1809-15.
3. Steen VD, Follansbee WP, Conte CG, Medsger TA, Jr. Thallium perfusion defects predict subsequent cardiac dysfunction in patients with systemic sclerosis. *Arthritis Rheum.* 1996;39(4):677-81.
4. Bulkley BH, Ridolfi RL, Salyer WR, Hutchins GM. Myocardial lesions of progressive systemic sclerosis. A cause of cardiac dysfunction. *Circulation.* 1976;53(3):483-90.
5. Kolto G, Faludi R, Aradi D, Bartos B, Kumanovics G, Minier T, et al. Impact of cardiac involvement on the risk of mortality among patients with systemic sclerosis: a 5-year follow-up of a single-center cohort. *Clin Rheumatol.* 2014;33(2):197-205.
6. Allanore Y, Wahbi K, Borderie D, Weber S, Kahan A, Meune C. N-terminal pro-brain natriuretic peptide in systemic sclerosis: a new cornerstone of cardiovascular assessment? *Ann Rheum Dis.* 2009;68(12):1885-9.
7. Ellis WW, Baer AN, Robertson RM, Pincus T, Kronenberg MW. Left ventricular dysfunction induced by cold exposure in patients with systemic sclerosis. *Am J Med.* 1986;80(3):385-92.
8. Fernandez-Codina A, Simeon-Aznar CP, Pinal-Fernandez I, Rodriguez-Palomares J, Pizzi MN, Hidalgo CE, et al. Cardiac involvement in systemic sclerosis: differences between clinical subsets and influence on survival. *Rheumatol Int.* 2017;37(1):75-84.
9. Mueller KA, Mueller II, Eppler D, Zuern CS, Seizer P, Kramer U, et al. Clinical and histopathological features of patients with systemic sclerosis undergoing endomyocardial biopsy. *PLoS One.* 2015;10(5):e0126707.
10. Follansbee WP, Zerbe TR, Medsger TA, Jr. Cardiac and skeletal muscle disease in systemic sclerosis (scleroderma): a high risk association. *Am Heart J.* 1993;125(1):194-203.
11. Hinchcliff M, Desai CS, Varga J, Shah SJ. Prevalence, prognosis, and factors associated with left ventricular diastolic dysfunction in systemic sclerosis. *Clin Exp Rheumatol.* 2012;30(2 Suppl 71):S30-7.
12. Maione S, Cuomo G, Giunta A, Tanturri de Horatio L, La Montagna G, Manguso F, et al. Echocardiographic alterations in systemic sclerosis: a longitudinal study. *Semin Arthritis Rheum.* 2005;34(5):721-7.
13. Mizuno R, Fujimoto S, Saito Y, Nakamura S. Cardiac Raynaud's phenomenon induced by cold provocation as a predictor of long-term left ventricular dysfunction and remodelling in systemic sclerosis: 7-year follow-up study. *Eur J Heart Fail.* 2010;12(3):268-75.
14. Saito M, Wright L, Negishi K, Dwyer N, Marwick TH. Mechanics and prognostic value of left and right ventricular dysfunction in patients with systemic sclerosis. *Eur Heart J Cardiovasc Imaging.* 2018;19(6):660-7.
15. Wangkaew S, Prasertwitayakij N, Phrommintikul A, Puntana S, Euathrongchit J. Causes of death, survival and risk factors of mortality in Thai patients with early systemic sclerosis: inception cohort study. *Rheumatol Int.* 2017;37(12):2087-94.
16. Chaosuwannakit N, Makarawate P. Value of cardiac magnetic resonance imaging in systemic sclerosis. *Reumatologia.* 2018;56(2):92-8.
17. Li X, Qian YQ, Liu N, Mu R, Zuo Y, Wang GC, et al. Survival rate, causes of death, and risk factors in systemic sclerosis: a large cohort study. *Clin Rheumatol.* 2018;37(11):3051-6.
18. Tennoe AH, Murbraech K, Andreassen JC, Fretheim H, Garen T, Gude E, et al. Left Ventricular Diastolic Dysfunction Predicts Mortality in Patients With Systemic Sclerosis. *J Am Coll Cardiol.* 2018;72(15):1804-13.
19. Eicher JC, Berthier S, Aho LS, Lorcerie B, Bonnotte B, Laurent G. Measurement of interatrial dyssynchrony using tissue Doppler imaging predicts functional capacity and cardiac involvement in systemic sclerosis. *Clin Exp Rheumatol.* 2014;32(6 Suppl 86):S-171-6.

20. Schioppo T, Artusi C, Ciavarella T, Ingegnoli F, Murgo A, Zeni S, et al. N-TproBNP as biomarker in systemic sclerosis. *Clin Rev Allergy Immunol.* 2012;43(3):292-301.
21. Kolto G, Vuolteenaho O, Szokodi I, Faludi R, Tornyos A, Ruskoaho H, et al. Prognostic value of N-terminal natriuretic peptides in systemic sclerosis: a single centre study. *Clin Exp Rheumatol.* 2014;32(6 Suppl 86):S-75-81.
22. Suzuki T, Ogasawara S, Ohsako-Higami S, Fukasawa C, Hara M, Kamatani N. Dipyridamole stress thallium perfusion scan for evaluating myocardial involvement in systemic sclerosis. *Mod Rheumatol.* 2001;11(3):210-6.
23. Steen VD, Medsger TA, Jr. Severe organ involvement in systemic sclerosis with diffuse scleroderma. *Arthritis Rheum.* 2000;43(11):2437-44.
24. Smith JW, Clements PJ, Levisman J, Furst D, Ross M. Echocardiographic features of progressive systemic sclerosis (PSS). Correlation with hemodynamic and postmortem studies. *Am J Med.* 1979;66(1):28-33.
25. Derk CT, Jimenez SA. Acute myocardial infarction in systemic sclerosis patients: a case series. *Clin Rheumatol.* 2007;26(6):965-8.
26. Sergiacomi G, De Nardo D, Capria A, Manenti G, Fabiano S, Borzi M, et al. Non-invasive diagnostic and functional evaluation of cardiac and pulmonary involvement in systemic sclerosis. *In Vivo.* 2004;18(2):229-35.
27. Kostis JB, Seibold JR, Turkevich D, Masi AT, Grau RG, Medsger TA, Jr., et al. Prognostic importance of cardiac arrhythmias in systemic sclerosis. *Am J Med.* 1988;84(6):1007-15.
28. Yiu KH, Schouffoer AA, Marsan NA, Ninaber MK, Stolk J, Vlieland TV, et al. Left ventricular dysfunction assessed by speckle-tracking strain analysis in patients with systemic sclerosis: relationship to functional capacity and ventricular arrhythmias. *Arthritis Rheum.* 2011;63(12):3969-78.
29. Rubio-Rivas M, Corbella X, Pestana-Fernandez M, Tolosa-Vilella C, Guillen-Del Castillo A, Colunga-Arguelles D, et al. First clinical symptom as a prognostic factor in systemic sclerosis: results of a retrospective nationwide cohort study. *Clin Rheumatol.* 2018;37(4):999-1009.
30. Coppi F, Giuggioli D, Spinella A, Colaci M, Lumetti F, Farinetti A, et al. Cardiac involvement in systemic sclerosis: identification of high-risk patient profiles in different patterns of clinical presentation. *J Cardiovasc Med (Hagerstown).* 2018;19(7):393-5.
31. De Luca G, Bosello SL, Gabrielli FA, Berardi G, Parisi F, Rucco M, et al. Prognostic Role of Ventricular Ectopic Beats in Systemic Sclerosis: A Prospective Cohort Study Shows ECG Indexes Predicting the Worse Outcome. *PLoS One.* 2016;11(4):e0153012.
32. Bernardo P, Conforti ML, Bellando-Randone S, Pieragnoli P, Blagojevic J, Kaloudi O, et al. Implantable cardioverter defibrillator prevents sudden cardiac death in systemic sclerosis. *J Rheumatol.* 2011;38(8):1617-21.
33. Chighizola C, Meroni PL, Schreiber BE, Coghlan JG, Denton CP, Ong VH. Role of N-terminal pro-brain natriuretic peptide in detecting clinically significant cardiac involvement in systemic sclerosis patients. *Clin Exp Rheumatol.* 2012;30(2 Suppl 71):S81-5.
34. D'Alto M, Cuomo G, Romeo E, Argiento P, Iudici M, Vettori S, et al. Tissue Doppler imaging in systemic sclerosis: a 3-year longitudinal study. *Semin Arthritis Rheum.* 2014;43(5):673-80.
35. Faludi R, Kolto G, Bartos B, Csima G, Czirjak L, Komocsi A. Five-year follow-up of left ventricular diastolic function in systemic sclerosis patients: determinants of mortality and disease progression. *Semin Arthritis Rheum.* 2014;44(2):220-7.
36. Bosello S, De Luca G, Berardi G, Canestrari G, de Waure C, Gabrielli FA, et al. Cardiac troponin T and NT-proBNP as diagnostic and prognostic biomarkers of primary cardiac involvement and disease severity in systemic sclerosis: A prospective study. *Eur J Intern Med.* 2019;60:46-53.